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A Case of Extra-axial Cavernous Hemangioma of the Cerebellar Falx

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Abstract

Extra-axial cavernous hemangioma (ECH) is clinically and radiologically distinct from parenchymal cavernous hemangioma. It usually arises intracranially with respect to the dura mater and most lesions arise from the middle cranial fossa, also known as cavernous sinus cavernous hemangioma. Few cases of extra-axial cavernous hemangioma arising from outside the middle cranial fossa have been reported, especially in the posterior cranial fossa. Here we report the rare case involving an adult male with cavernous hemangioma in the cerebellar falx and examine both imaging and symptom features as they relate to diagnosis and treatment, taking into consideration previous research findings.

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Intracranial cavernous hemangioma occurs in 0.39-0.90% of the population as estimated from magnetic resonance imaging (MRI) studies,[1] and is thought to account for 3-15% of all intracranial vascular malformations [2,3]. Cavernous hemangioma most frequently occurs in the subcortical white matter of the cerebral hemispheres and less commonly in locations including the cerebellopontine angle, pineal gland, and ventricles [1,4-9]. Few cases of primary extra-axial cavernous hemangioma have been reported. Of the reported cases, almost all describe hemangioma arising from the cavernous sinus of the middle cranial fossa [4,5,8,10,11]. Instances of extra-axial cavernous hemangioma located outside of the middle cranial fossa are very rare. This report describes a rare case of dural cavernous hemangioma arising from the cerebellar falx.

Case Report

A 56-year-old right-handed male in excellent physical health developed dysarthria and right-hand ataxia that gradually worsened over the course of 6 months. He was referred to our hospital after developing gait disturbance. Upon admission, the patient presented with ataxic dysarthria, poor tandem gait, and poor pointing performance in the finger nose test. He was assessed with a modified Rankin scale score of 3.

Radiological Findings

Computed tomography (CT) demonstrated a large, extra-axial mass with no calcification attached to the cerebellar falx in the right cerebellar hemisphere. MRI revealed an extra-axial mass measuring 55 mm that was isointense with the surrounding gray matter on T1-weighted images and hyperintense on T2-weighted images. The mass showed intense and homogeneous enhancement after gadolinium-diethylenetriamine penta-acetic acid (Gd-DTPA) administration. The mass had a dural attachment at the base of the lesion measuring 1 cm, but there was no evidence of an enhanced dural tail. The lesion compressed the fourth ventricle, blocking the flow of CSF and causing hydrocephalus. Angiography did not demonstrate any tumor staining (Figure 1). Accordingly, the patient was preoperatively diagnosed with extra-axial hemangioma or meningioma.

Operative Findings

We elected to perform a suboccipital craniotomy. After opening the dura, we identified a tumor in contact with the right cerebellar hemisphere attached to the cerebellar falx measuring 1 cm. The adhesion to the falx was very strong and hemorrhagic (Figure 2A). We used bipolar forceps to stop the bleeding and begin cutting away the attachment of the tumor to the falx. The removed mass resembled a stiff, reddish-brown, blood-filled sponge and differed in appearance from the usual presentation of cavernous hemangioma near the brainstem (Figure 2B). There were no vascular attachments to the brain surface or another dura, and there was preservation of the arachnoid plane. We performed a total en bloc resection with minimal blood loss and preservation of the dura.

Histopathological Findings

Hematoxylin and eosin staining showed typical cavernous hemangioma with no distinguishable differences from lesions typically occurring in the parenchyma. In contrast, the tissue around the dural attachment was composed of fiber components. Tumor cells lining the vascular channels were positive for CD34 but negative for glia fibrillary acidic protein (GFAP) and epithelial membrane antigen (EMA). We did not detect any meningothelial cells, and intervascular substances were hyalinized and hypocellular, excluding a diagnosis of meningioma (Figure 3).

Postoperative Course

After the operation, we observed rapid improvement in the patient's hydrocephalus and overall condition. There were no signs of recurrence 1 year later.

Discussion

Cavernous hemangioma is a vascular malformation that can occur anywhere in the central nervous system; however, it is usually detected in the subcortical white matter of the cerebral hemispheres. Extra-axial cavernous hemangioma is rare and exclusively located outside

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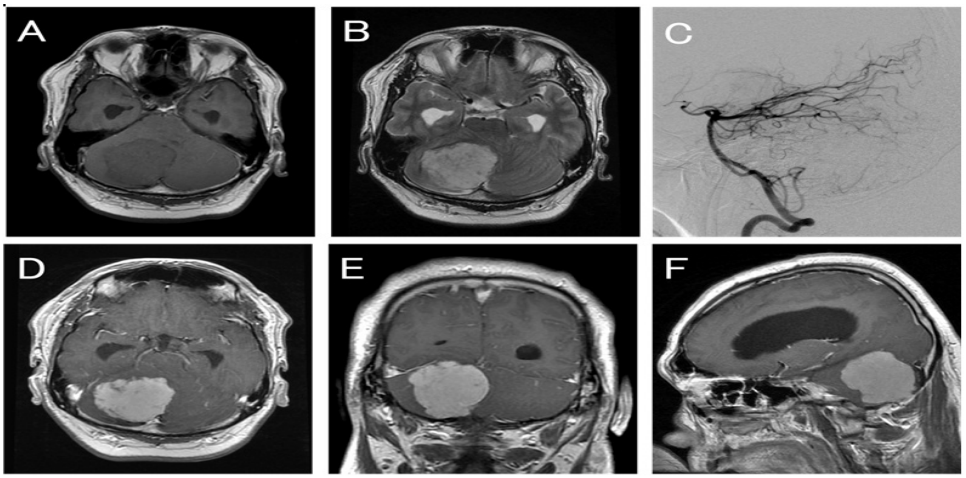

Figure 1: Magnetic resonance imaging scans revealing the extra-axial mass measuring 55 mm with isointensity on T1-weighted images (A) and hyperintensity on T2-weighted images (B). Angiography demonstrating the absence of tumor stains (C). Homogeneous enhancement of the lesion after gadolinium injection (D, E, F).

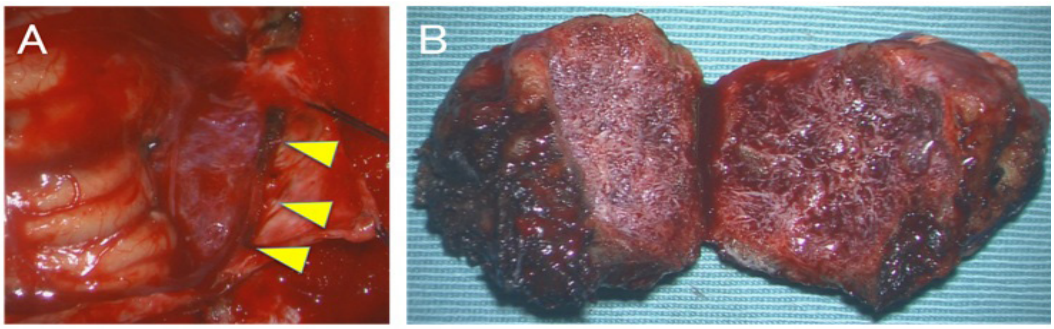

Figure 2: Intraoperative photograph showing the lesion attached to the cerebellar falx [arrow heads] (A). Photograph of the en bloc resected mass resembling a stiff reddish-brown blood-filled sponge (B).

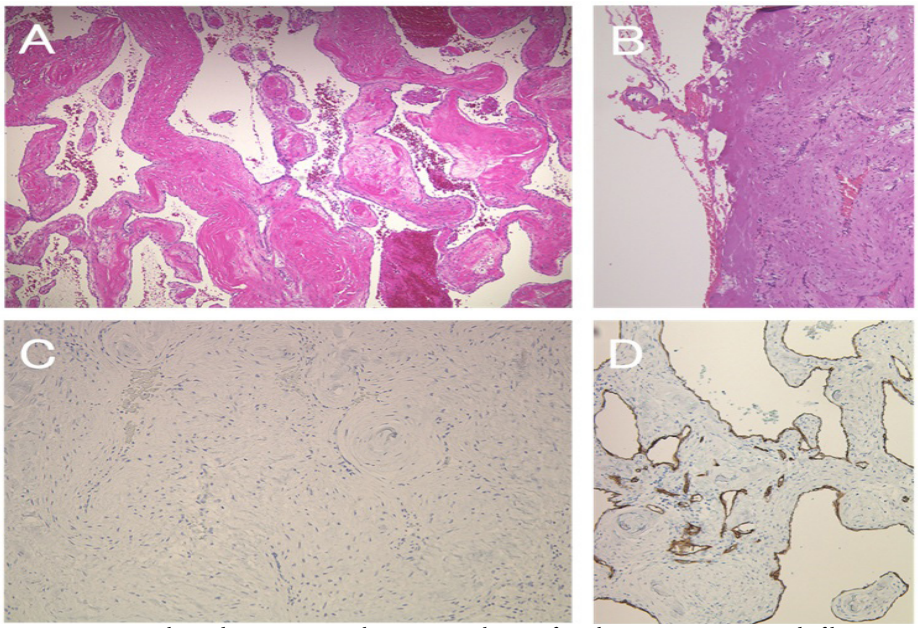

Figure 3: Hematoxylin and eosin staining showing typical spongiform hemangioma comprised of large vascular channels, the external limits of which were on the dura mater and composed of fiber components (A, B). The lesion was negative for EMA (C) and positive for CD34 (D).

of the middle cranial fossa, with only 15 reported cases of extra-axial cavernous hemangioma in the posterior cranial fossa [1]. Lesions in the middle cranial fossa are predominately found in women. In contrast, lesions in the posterior cranial fossa are more frequent in men [1,11], as in our case. Among cases in the posterior fossa, the tentorium cerebelli is the most common site; only two cases arising from the cerebellar falx have been reported previously [1,12].

Cavernous hemangioma is often asymptomatic, and when symptoms are present they depend on the location and size of the lesion. Clinical manifestations of intra-axial cases, the most important of which are epilepsy and focal neurological deficits, are due to compression with multiple episodes of small parenchymal hemorrhage and lesion enlargement [4-9]. Conversely, extra-axial cavernous hemangioma leads to seizures in only 22% of cases, while 75% of extra-axial lesions located outside of the middle cranial fossa produce headache [13]. Large tumors tend to cause headache whereas small lesions can induce episodes of severe headache, potentially as the result of dural irritation [13]. Focal neurological deficits due to compression are also a major symptom of extra-axial lesions [8,10,13]. In our case, an extra-axial lesion was associated with cerebellar ataxia and obstructive hydrocephalus, but no headache or epilepsy.

Extra-axial cavernous hemangioma appears on CT as a hyperdense or heterogeneous mass with no calcification. On MRI, lesions are hypointense or isointense on T1-weighted images and hyperintense on T2-weighted images. The use of a contrast medium yields homogenous enhancement and permits the visualization of lesion attachments to the dura. Some reports have described the presence of the dural tail sign in cases of meningioma [4]. Yet, a dural tail can be visualized in cases of hemangioma as the result of a meningeal reaction or a direct tumor extension, leading to misdiagnosis as meningioma [14]. Accordingly, the absence of hemosiderosis indicating a history of bleeding in extra-axial cavernous hemangioma often leads to an incorrect preoperative diagnosis of meningioma. In a study sponsored by the National Cancer Institute, enhanced CT accurately detected 96.2% of intracranial meningiomas [2]. Yet, more than half of lesions that were histologically diagnosed as extra-axial cavernous hemangioma were preoperatively misdiagnosed as meningioma. Our case did not exhibit the dural tail sign or hemosiderosis. Therefore, we were unable to exclude a diagnosis of meningioma preoperatively. Even in the absence of hemosiderosis, cavernous hemangioma should be included in the differential diagnosis of dural lesions [1,4].

In general, surgical treatment is recommended for extra-axial cavernous hemangioma when patients report headache or neurological deficits. In such cases, the location of the mass and the difficulty of surgical resection are important factors. Lesions in the middle cranial fossa are clinically aggressive and difficult to resect because they grow towards the cavernous sinus and parasellar region [15]. These lesions also have wide attachments with the dura mater and are hemorrhagic, with a hypervascular appearance in the middle arterial phase to the late venous phase on angiography [9]. As such, total resection is often difficult and reported rates of preoperative mortality and morbidity are significantly higher. Recently, preoperative radiosurgery or embolization was proposed to reduce bleeding prior to resection of extra-axial cavernous hemangioma [1]. In contrast, lesions in other locations have an avascular appearance given slow flow and are easily and successfully resected with minimal blood loss [2-4,8,10,11,13,15]. In our case, surgery was completed safely with minimal blood loss in the absence of any pretreatment.

Conclusion

Extra-axial cavernous hemangioma in the posterior cranial fossa and especially lesions attached to the cerebellar falx are quite rare. These lesions can closely resemble meningioma on radiological imaging; therefore, the differential diagnosis of extra-axial lesions should include both meningioma and hemangioma. Finally, the location of the lesion is an important factor for predicting the surgical treatment outcome. Extra-axial cavernous hemangioma can be successfully removed without preoperative radiosurgery or embolization, except for lesions in the middle cranial fossa.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

FS and YK designed the case report and oversaw its direction. FS wrote the manuscript, designed the figures with input from all authors, and assumed responsibility for the integrity of the data and accuracy of the analysis. YW aided in analyzing histological findings and drafting of the manuscript. HO supervised the case study. FS, YK, KT, YK, SY, KT, RM, TM, YW, and HO discussed the results and reviewed the manuscript for important intellectual content. All authors have approved the version submitted for publication.

Ethical considerations

The patient gave us his written informed consent for publication of this case report and accompanying images.

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References

1. Ito M, Kamiyama H, Nakamura T, Nakajima H, Tokugawa J, et al. (2009) Dural cavernous hemangioma of the cerebellar falx. Neurol Med Chir (Tokyo) 49: 410-412.
2. Perry JR, Tucker WS, Chui M, Bilbao JM (1993) Dural cavernous hemangioma: an under-recognized lesion mimicking meningioma. Can J Neurol Sci 20: 230-233.
3. Vogler R, Castillo M (1995) Dural cavernous angioma: MR features. AJNR Am J Neuroradiol 16: 773-775.
4. Sakakibara Y, Taguchi Y, Nakamura H, Matsumori T, Wakui D, et al. (2015) Dural cavernous angioma: a case report and review of the literature. No Shinkei Geka 43: 429-434.
5. Batra S, Lin D, Recinos PF, Zhang J, Rigamonti D, et al. (2009) Cavernous malformations: natural history, diagnosis and treatment. Nat Rev Neurol 5: 659-670.
6. Farmer JP, Cosgrove GR, Villemure JG, Meagher-Villemure K, Tampieri D, et al. (1988) Intracerebral cavernous angiomas. Neurology 38: 1699-1704.
7. Gross BA, Lin N, Du R, Day AL (2011) The natural history of intracranial cavernous malformations. Neurosurg Focus 30: 24.
8. Requena I, Arias M, López-Ibor L, Pereiro I, Barba A, et al. (1991) Cavernomas of the central nervous system: clinical and neuroimaging manifestations in 47 patients. J Neurol Neurosurg Psychiatry 54: 590-594.
9. Sakakibara Y, Taguchi Y, Uchida K (2010) A case of cavernous angioma presenting as migrainous attack. No Shinkei Geka 38: 287-291.

10. Biondi A, Clemenceau S, Dormont D, Deladoeuille M, Ricciardi GK, et al. (2002) Intracranial extra-axial cavernous (HEM) angiomas: tumors or vascular malformations? J Neuroradiol 29: 91-104.

11. Sohn CH, Kim SP, Kim IM, Lee JH, Lee HK, et al. (2003) Characteristic MR imaging findings of cavernous hemangiomas in the cavernous sinus. AJNR Am J Neuroradiol 24: 1148-1151.

12. Melone AG, Delfinis CP, Passacantilli E, Lenzi J, Santoro A, et al. (2010) Intracranial extra-axial cavernous angioma of the cerebellar falx. World Neurosurg 74: 501-504.

13. Shen WC, Chenn CA, Hsue CT, Lin TY (2000) Dural cavernous angioma mimicking a meningioma and causing facial pain. J Neuroimaging 10: 183-185.

14. Gupta RK, Saran RK, Jagetia A, Narang P (2016) Extra-axial dural cavernous hemangioma with dural tail sign, masquerading as meningioma. J Neurosci Rural Pract 7: 615-616.

15. Lewis AI, Tew JM Jr, Payner TD, Yeh HS (1994) Dural cavernous angiomas outside the middle cranial fossa: a report of two cases. Neurosurgery 35: 498-504.